### Case Report

DOI: http://dx.doi.org/10.18203/issn.2454-5929.ijohns20180737

## Gradinego's syndrome: atypical presentation

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Received: 08 January 2018 Revised: 08 February 2018 Accepted: 10 February 2018

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#### **ABSTRACT**

Gradenigo's syndrome was first described by Giuseppe Gradenigo in 1907 when he reported a triad of symptoms consisting of unilateral periorbital pain related to trigeminal nerve involvement, diplopia due to sixth cranial nerve palsy and persistent otorrhoea, associated with otitis media with petrositis. The classical syndrome related to otitis media has become very rare after the antibiotic era. Incomplete and atypical clinical features of Gradenigo's syndrome have been described and non infectious causes may mimic this condition. We report a case of acute petrositis in a 19 year old boy with unilateral periorbital pain, diplopia (lateral rectus palsy) in the absence of ear discharge. Careful clinical history, physical examination, including neuroimaging, is necessary to make a differential diagnosis. Appropriate management requires antibiotic treatment and possible surgical intervention.

**Keywords:** Gradinego's syndrome, Petrositis, Lateral rectus palsy

#### **INTRODUCTION**

Gradenigo's syndrome was first described in 1907 by Giuseppe Gradenigo, when he reported a triad of symptoms characterised by unilateral periorbital pain due to trigeminal nerve involvement at Meckel's cave, diplopia due to involvement of abducens nerve in Dorello's canal and persistent otorrhoea of the involved ear. The classical triad is very rare in this era of antibiotics. The incidence of apical petrositis is now reportedly two per 100,000 children with acute otitis media.<sup>2</sup> Gradenigo's syndrome is often recognized late due to subtlety of its signs and symptoms.<sup>3</sup> The complete triad may not be often present, but can develop if the condition is not treated correctly. We present a case of an atypical presentation of Gradenigo's syndrome in an adolescent boy with unilateral periorbital pain, diplopia (lateral rectus palsy) in the absence of ear discharge.

#### **CASE REPORT**

A 19 year old boy presented with the history of left ear pain for 2 weeks, fever with chills and rigor since 10 days following an episode of upper respiratory tract infection. He had history of double vision, pain in the left eye and restricted movement of eye towards left since 1 week. He also had headache over the left temporal region since 1 week. But he did not have ear discharge, decreased hearing, nausea or vomiting. On examination, his vitals and systemic examination were normal. On examination of his left ear, there was loss of cone of light in the tympanic membrane, bulge in the pars tensa and increased vascularity along handle of malleus (Figure 1). Rest of the ENT examination was normal.

Examination of eye revealed diplopia in left lateral gaze and there was left lateral rectus palsy (Figure 2).

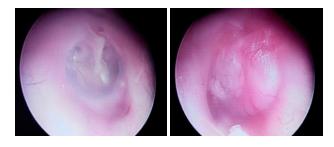


Figure 1: Otoendoscopic picture of right and left tympanic membrane.



Figure 2: A=Forward gaze; B=Right lateral gaze; C=Left lateral gaze.

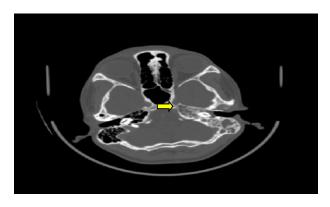


Figure 3: CT Brain Plain showing left otomastoiditis with petrous apicitis (arrow).



Figure 4: CT Brain with contrast showing enhancing left abducent nerve and enhancing soft tissue thickening in the region of left Meckel's cave (arrow).

On investigation, his routine blood investigations were normal. His audiogram showed a mild conductive hearing loss on left side with a type "B" curve on tymponametry.

CT brain, plain and contrast was done. There was complete soft tissue opacification of left mastoid air cells, middle ear cavity and left petrous apex, suggestive of left otomastoiditis with petrous apicitis (Figure 3). There was a thin linear enhancing structure extending anterolaterally from left pontomedullary junction- enhancing left abducent nerve. Enhancing soft tissue thickening was seen in the region of left Meckel's cave (Figure 4).

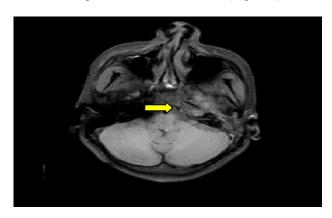


Figure 5: MRI Brain plain T1W showing iso-intensity at left petrous apex (arrow).

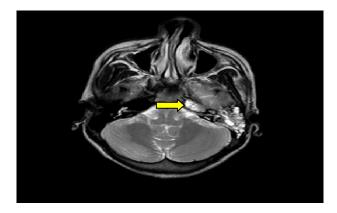


Figure 6: MRI Brain T2W showing hyperintensity at left petrous apex (arrow) and mastoid air cells.

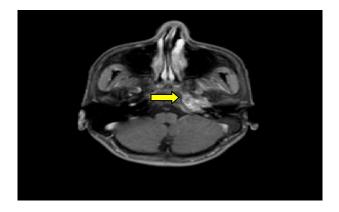


Figure 7: MRI Brain post contrast T1W showing enhancing dural/soft tissue thickening in the region of clivus, Meckel's cave with possible compression of left abducent nerve (arrow).

MRI brain with contrast showed iso to hyperintensity in T1 and hyperintensity at the left petrous apex and mastoid air cells in T2 (Figure 5 and 6) suggesting features of left petrous apicitis and otomastoiditis with enhancing dural/soft tissue thickening in the region of clivus, Meckel's cave with possible compression of left abducent nerve (Figure 7). There was no definite cortical venous/ sinus thrombosis.

He was started on IV antibiotics- Injection Ceftriaxone 1 g Q12H, Inj Amikacin 500 mg Q24H for a week. He was also given oral analgesics and nasal decongestant drops for one week. After a week of IV antibiotics, he was planned for mastoid exploration under general anaesthesia. Cortical mastoidectomy was done. There was hypertrophied mucosa in the antrum and aditus, which was removed and aditus patency was established. Ossicular chain was found to be intact. There was no pus or cholesteatoma sac seen.

Neurology/Neurosurgery opinion was sought. Considering risk/benefit with surgical approach to petrous apex, long course IV antibiotic was suggested. He was treated with a total of 7 weeks of IV Ceftriaxone 1 g Q12H.

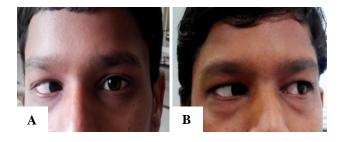


Figure 8 (A and B): Left lateral gaze- pre and post treatment.

His left ear pain and retrobulbar pain reduced in 1 week. There was improvement in abduction of left eye in 1 week. Complete resolution of left lateral rectus palsy was seen after 7 weeks of IV antibiotics (Figure 8).

#### **DISCUSSION**

Petrous apicitis, also known as Gradenigo's syndrome, is a non localized infection with inflammatory changes involving the mucosa and bone of apex of petrous temporal bone with similar findings in the middle ear cleft. Petrous apex which lies between the inner ear and clivus, forms the medial part of temporal bone. It is divided into 2 compartments: anterior and posterior, by the internal auditory canal. The larger anterior part of the petrous apex is more frequently involved with disease processes. The petrous bone may be pneumatized (filled with mucosa-lined air cells), diploic (filled with bone marrow), or sclerotic. When pneumatized, the pattern of pneumatization is quite symmetrical. Pneumatization of petrous apex is seen in about 30% of adults.

There are various pathways for spread of infection by cell tracts from mastoid to petrous apex. These are posterosuperior, posteromedial, sub-arcuate, peritubal and perilabyrinthine. Posterosuperior tract starts in the mastoid and runs behind or above the bony labyrinth to the petrous apex, subarcuate tract passes through the arch of superior semicircular canal to reach the apex. Infective process runs along these cell tracts to reach the petrous apex. The trigeminal ganglion and the sixth cranial nerve are separated from the bony petrous apex only by the dura mater, hence their vulnerability to inflammatory processes occurring within this region.<sup>6</sup>

Dorello described an osteofibrous canal (Dorello's Canal), consisting of abducent nerve and inferior petrosal sinus, at the tip of the pars petrosa temporalis. Abducent nerve gets affected in this canal by inflammation. 8,9

The spread of infection can occur through pneumatization, vascular channel or bone erosion. 9-11 It can be seen as a complication of acute or chronic otitis media. In our case, history and clinical findings are more consistent with acute otitis media as there was no ear discharge and tympanic membrane was intact.

There is no statistical data about the age distribution and the type of onset (acute versus chronic) of Gradenigo's syndrome. However, considering the literature, chronic otitis media seems to be the main leading cause in adults, whereas acute otitis media without ear discharge is more common in the pediatric age group. <sup>9,10</sup> An adult patient having acute otitis media presenting with the acute form of a petrous apicitis was reported by Ulkamen. <sup>12</sup> But we had an adolescent patient presenting with the syndrome following acute otitis media.

Before the antibiotic era, petrous apicitis was associated with severe complications like labyrinthitis, meningitis, intracranial abscess formation, retropharyngeal abscess formation, venous sinus thrombosis, cranial nerve palsies.<sup>13</sup>

Deep pain in the ear and retrobulbar region is the most common symptom in Gradenigo's syndrome, as in our case.<sup>5</sup>

Many patients do not present with all the three components of the syndrome. In 57 cases of petrous apicitis reported by Gradinego, 24 cases had the pure triad, 29 of them had additional cranial nerve deficits, and 4 cases had meningitis resulting in death.<sup>5</sup> Whereas, among 8 cases of petrous apecitis described by Chole and Donald, only 1 case had pure triad.<sup>5</sup>

Gradenigo's syndrome has been mostly treated by radical surgery, but there are some reports of conservative management. Marianowski et al reported a case of a 6-year-old child, who was completely cured by myringotomy and antibiotic therapy. Burston et al reported two cases of Gradenigo's syndrome, one in a 70-

year-old man and the other in a 6-year-old child, who were also managed conservatively without the need for radical surgery. Similarly, Marteau et al achieved total cure in a 4 year old girl by only antimicrobial treatment. In another case reported by Ulkamen et al, a 56 year old man, was managed with myringotomy with ventilation tube insertion and 10 days of intravenous ceftriaxone followed by oral antibiotic therapy. Al-Ammar also reported a case of Gradenigo's syndrome managed with myringotomy and ventilation tube insertion, with a good outcome, but still had recurrent symptoms of the syndrome after the extrusion of the ventilation tube. The remaining reported cases were generally treated by radical surgery.

In our case, diagnosis of Gradenigo's syndrome was made based on history, clinical findings and neuro-imaging which showed T2 hyperintensity in petrous apex, enhancing dural/soft tissue thickening in the region of clivus, Meckel's cave with possible compression of left abducent nerve. Prompt treatment with IV antibiotics and adequate mastoid drainage led to the recovery of symptoms and lateral rectus palsy preventing intracranial complications.

Funding: No funding sources Conflict of interest: None declared Ethical approval: Not required

#### **REFERENCES**

- 1. Gradenigo G. Ueber die paralyse des Nervus abducens bei Otitis. Arch Ohrenheilunde. 1907;774:149–87.
- 2. Goldstein NA, Casselbrant ML, Bluestone CD, Kurs-Lasky M. Intratemporal complications of acute otitis media in infants and children. Otolaryngol Head Neck Surg. 1998;119:444–54.
- 3. Tutuncuoglu S, Uran N, Kavas I, Ozgur T. Gradenigo syndrome: a case report. Pediatr Radiol. 1993;23:556.
- 4. Jackler RK, Parker DA. Radiographic differential diagnosis of petrous apex lesions. Am J Otolaryngol. 1992;13:561–74.

- 5. Chole RA. Petrous apicitis: surgical anatomy. Ann Otol Rhinol Laryngol. 1985;94:251–7.
- Kantas I, Papadopoulou A, Balatsouras DG, Aspris A, Marangos N. Therapeutic approach to Gradenigo's syndrome: a case report. J Med Case Rep. 2010;4:151.
- 7. Felisati D, Sperati G. Gradenigo's syndrome and Dorello's canal. Acta Otorhinolaryngol Ital. 2009;29(3):169-72.
- 8. Gibier L, Darrouzet V, Franco-Vidal V. Gradenigo syndrome without acute otitis media. Pediatr Neurol. 2009;41(3):215-9.
- 9. Marianowski R, Rocton S, Ait-Amer JL, Morisseau-Durand MP, Manach Y. Conservative management of Gradenigo syndrome in a child. Int J Pediatr Otorhinolaryngol. 2001;57(1):79-83.
- 10. Burston BJ, Pretorius PM, Ramsden JD. Gradenigo's syndrome: successful conservative treatment in adult and paediatric patients. J Laryngol Otol. 2005;119:325-9.
- 11. Gillanders DA. Gradenigo's syndrome revisited. J Otolaryngol. 1983;12:169-74.
- 12. Ulkumen B, Kaplan Y. Conservative treatment of Gradenigo's syndrome triggered by acute otitis media. Pak J Med Sci. 2012;28(4):735-7.
- 13. Hardjasudarma M, Edwards RL, Ganley JP. Magnetic resonance imaging features of Gradenigo's syndrome. Am J Otolaryngol. 1995;16:247-50.
- 14. Marteau E, Georget-Bouquinet E, Verlhac S, Gauthier A, Remus N, Madhi F. Successful prolonged conservative treatment of Gradenigo's syndrome in a 4-year-old girl: A case report and literature review. Int J Pediatr Otorhinolaryngol Extra. 2011;6(2):100-3.
- 15. Al-Ammar AY. Recurrent temporal petrositis. J Laryngol Otol. 2001;115:316-8.

Cite this article as: Sumana CV, Hasan S. Gradinego's syndrome: atypical presentation. Int J Otorhinolaryngol Head Neck Surg 2018;4:604-7.